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Coexistance of Pemphigus Vulgaris and Psoriasis in a Middle Aged Man

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Abstract

Psoriasis and pemphigus vulgaris are two inflammatory and autoimmune diseases. Their concomitant occurrence in one patient is very rare. We describe a 57-year-old patient with a history of severe plaque type psoriasis for 2 years. After a few months, he presented with disseminated bullae and mucosal erosions which were documented as pemphigus vulgaris. Both of the diseases were successfully controlled with rituximab, prednisolone and weekly methotrexate.

Keywords: Psoriasis, Pemphigus Vulgaris, Rituximab

Introduction

The coexistence of psoriasis vulgaris and bullous diseases has been described in less than 100 literature[1-14]. The triggering factor in association of psoriasis and pemphigus vulgaris is unknown and may be more than coincidence and may be explained on an immunologic basis. The effects of one immune-mediated skin disease may influence the occurrence of the other one. Possible mechanisms include reduced barrier function of the skin and antigen modification, altered regulation of T-cell activity, plasminogen activator abnormalities and side effects of therapies. The concept of 'epitope spreading' by exposing new antigens due to cutaneous damage may explain development of bullous disorders. Psoriasis also has been associated with other autoimmune diseases like crohn, lupus erythematosus, rumatoidartritis and celiac disease[15]. Methotrexate is first-line systemic therapy for psoriasis as it is highly efficacious for severe disease and all clinical variant of psoriasis. In some patients with pemphigus vulgaris, methotrexate has been used as an immunosuppressive agent. Also rituximab increasingly is used for pemphigus vulgaris. Using as adjuvant therapy led to a complete remission in most of the patients with refractory pemphigus vulgaris[16,17]. Concomitant use of methotrexate and rituximab is approved in patients with rheumatoid arthritis. In the literature, the coexistence of psoriasis vulgaris and bullous diseases has been treated with cyclosporine monotherapy[2,3], etanercept[1], corticosteroids and cyclophosphamide[4], corticosteroid and azathioprine[5], corticosteroid monotherapy, methotrexate monotherapy. We present a patient with a history of severe plaque type psoriasis treated with Neotigasone that developed flaccid skin and mucosal bullae documented as pemphigus vulgaris by histopathology and direct immunofluorescence. Both of the diseases were successfully controlled by rituximab 500 mg/ IV/ weekly for 4 weeks, prednisolone daily and methotrexate weekly. Maybe immunosuppressive agents directed at cellular and humoral responses can result in clinical improvement of both conditions.

Case Report

A 57-year-old man presented to the Dermatology clinic of Sina hospital in July. 2015, with a 2-month history of erosions and crusted lesions and flaccid bullae of the oral mucosa and skin. These lesions began to erupt on his oral mucosa with eventual spread to the scalp, face, limbs include palmoplantar skin (Figure 1-3). In physical exam his Nickolsky sign was also positive. His Past medical history was included plaque type psoriasis from 2 years ago. His drug history was included oral Neotigasone (30 mg/day) and topical corticosteroid. He was receiving Neotigasone from 9 month ago. On presentation to our clinic, biopsy was done. Histology and direct immunofluorescence assay confirmed diagnose of pemphigus vulgaris. He was admitted in dermatology unit. Prior to treatment, chest X-ray,

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PPD (purified protein derivate skin test), CBC, liver function tests, renal function tests, FBS, lipid profile, hepatitis B and C serologic profiles were done. All lab data was normal. Patient was received rituximab 500 mg/ IV/ weekly for 4 weeks, prednisolone daily and methotrexate weekly.

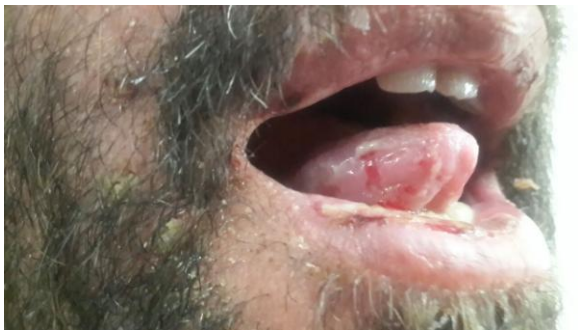

Figure 1. Tongue, lip and buccal mucosae erosion

Figures 2. disseminated flaccid bullae and erosionon trunkand limbs

Methotrexate was started with dose of 5 mg/ weekly and increased by 2.5 mg/ weekly and then continue with dose of 12.5 mg/ weekly. Prednisolone was started with dose of 75mg/ daily, then gradually tapered over 5 months, and reached maintenance dose of 7.5 mg/ daily. At this time, we checked IgG autoantibody against desmoglein 3 and desmoglein 1 by ELISA, both of them were negative and below 20 (anti Dsg1:0.2 and anti Dsg3:0.7). Now, the patient is in remission and No flare-up (neither psoriasis lesions nor pemphigus lesions) have been observed after 12 months treatment.

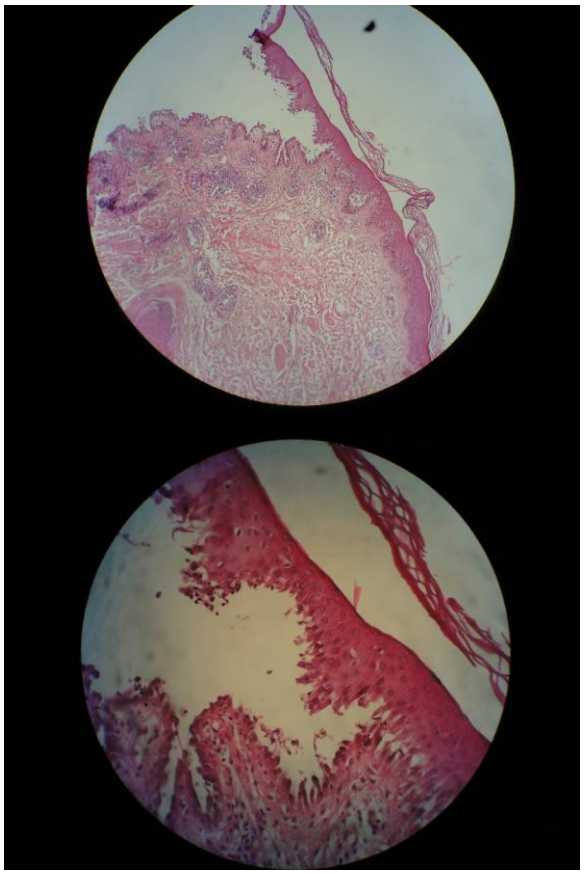

Figure 4. (A) Blister in the skin shows suprabasilaracantholysis, papillae protrude into the blister cavity and there is moderate perivascular infiltration. (B)Few rounded up acantholytic cells are seen in blister cavity, attachment ofbasal cells to the basement membrane has made the appearance of 'row of tombstones'.

Discussion

For the past two decades, psoriasis has been regarded as a T cell driven disease. Recent studies have shown that the role of Th17 in its pathogenicity has also a role in autoimmunity; however, to date no true autoantigen has been definitively identified [15].

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Pemphigus vulgaris is an autoimmune blistering disease that is characterized by finding of IgG autoantibody against desmoglein 3 and desmoglein 1.

We treated our patient with prednisolone, methotrexate and rituximab. The diseases were successfully controlled by rituximab 500 mg/ IV/ weekly for 4 weeks, prednisolone daily and methotrexate weekly. It seems that immunosuppressive agents directed at cellular and humoral responses can result in clinical improvement of both conditions.

Conflicts of interest

Authors declare no conflict of interests.

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